a manifestation of lymphoma, and all died before receiving chemotherapy. If a patient with WM develops ascites, clinicians should consider the possibility that the disease has transformed into IL, especially if the patient has received alkylating agents.

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Acute Leg Ischaemia as a Presentation of Hyperleukocytosis Syndrome in Acute Myeloid Leukaemia

To the Editor: Recently, Campbell and Mitchell [1] reported the first case of acute leg ischaemia as a presentation of hyperleukocytosis syndrome in acute myeloid leukaemia (AML). We present a similar case in a patient without previous clinical ischaemia.

In 1985 a 42-year-old smoker was admitted for pain in his right leg which had lasted 2 days. Examination revealed absent tibial and pedal pulses. Full blood examination revealed haemoglobin 9.8 g/dl, platelets 30×10^9/l, white cell count (WCC) 150×10^9/l (95% blasts). The coagulation study was normal. Bone marrow examination confirmed the diagnosis of AML-M2. Angiography demonstrated thrombosis on the second sector of the right popliteal artery. Arterial thrombolysis with streptokinase was performed without success, and a cytoreduction with hydroxyurea (8 g/day) was later performed. Following the hydroxyurea therapy, WCC dropped to 60×109/l. After three thrombectomies in 6 days, with subsequent stenosis in all cases, treatment with cytosine arabinoside, daunorubicin, and thioguanine was begun; in spite of this, necrotic signs in the right leg were evident and right supracondylar amputation was decided upon. After standard chemotherapy for AML complete remission was reached. Two years later a bone marrow transplant was performed, but the patient died on day 17 following the transplant, as a consequence of a diffuse alveolar haemorrhage that was confirmed in the postmortem examination.

The presence of arterial leukocyte thrombi was clearly demonstrated in the autopsy analysis, especially in a patient affected by AML with affectation fundamentally at the level of small calibre vessels [2]. This association is more evident when hyperleukocytosis [3] or previous arterial damage [4] exist. Though major vessel obstructions are uncommon, we believe that this is probably based on underestimates; our case data of 1985 and the reading of the Campbell and Mitchell case [1] encouraged us to revise our file of leukaemias, which induced us to think that unpublished cases could exist. Campbell and Mitchell propose that developed hyperleukocytosis produced leukostasis at the site of an atherosclerotic stenosis. We report the second case of peripheral arterial thrombosis where the age of the patient, the absence of symptoms and signs, and the nonappearance of atherosclerosis in the autopsy lead us to believe that the increase in the secondary sanguine viscosity to the hyperleukocytosis was sufficient to produce vascular occlusion.

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Simultaneous Occurrence of Lupus Anticoagulant, Factor VIII Inhibitor and Localized Pemphigoid

To the Editor: Acquired hemophilia, a rare disorder, has been observed in association with various autoimmune diseases [1]. Among them, bullous skin lesions have been reported, but rarely bullous pemphigoid (BP) [2]. Lupus anticoagulant (LA) is also encountered in many clinical states characterized by immunologic disorders [3]. Herein, we describe a case of localized pemphigoid associated with both LA and factor VIII inhibitor. To our knowledge, this combination has not yet been reported.

A 92-year-old woman, 2 weeks after apparition of a bullous dermatosis on the left leg, developed a large ecchymosis over the upper extremities and abdomen. She had no family or past history of a bleeding disorder. Histological and immunofluorescence studies confirmed the diagnosis of BP. Blood cell count, platelet count, and prothrombin time were within normal ranges. Activated partial thromboplastin time (APTT) was prolonged (102 sec; control value: 34 sec). The addition of normal plasma (1:1) failed to correct the APTT, leading us to suspect an acquired anticoagulant. The presence of LA was confirmed by a neutralization procedure using hexagonal (II) phase phosphatidylethanol (Staclot LA, Diagnostica Stago, Asnieres, France). The severity of bleeding prompted us to suspect an associated clotting factor deficiency. Fibrinogen factors II, V, IX, XI, and XII were within normal ranges, but factor VIII level was 2%. The presence of a factor VIII inhibitor was confirmed using the Bethesda method [4]. An antihuman factor VIII inhibitor of 32 Bethesda units (BU) was identified while the titer of antiporcine VIII inhibitor activity was 2 BU. Antinuclear antibodies, antidouble-stranded DNA antibodies, and anticardiolipin antibodies were not detected.

Acute bleeding was controlled using porcine VIIIC: HyateC (Speywood, Berkshire, UK) at 50 U/kg, three times a day. No recurrent bleeding was observed during the hospitalization. Treatment was disrupted after 3 days, while a prednisolone therapy (1 mg/kg/day) was initiated. After 2 weeks of steroid therapy, dermatological lesions disappeared, while biological abnormalities were less pronounced: factor VIII increased to 15%, with an